

Case Report

An unusual cervical swelling in a child

Ahmed H. Al-Salem, FRCSI, FACS.

ABSTRACT

Congenital thymic cysts are rare benign lesions, frequently seen in the neck and mediastinum, but because of their infrequent occurrence and similarity to other more common cystic swellings, their preoperative diagnosis is not considered. Congenital thymic cysts although rare should be considered in the differential diagnosis of cervical and mediastinal cystic swellings in children. This is a case report of a multilocular cervical thymic cyst in a child.

Keywords: Thymus, cyst.

Saudi Med J 2001; Vol. 22 (10): 917-919

The thymus gland develops from the 3rd pharyngeal pouch in the neck and descends down as thymopharyngeal tracts to the final position in the superior mediastinum.¹ As a result, ectopic thymic tissue may be found along the line of descent from the original position in the neck to the final position in the superior mediastinum, and although it is not uncommon to find ectopic thymic tissue during autopsy studies or neck exploration for other reasons, very few enlarge to present clinically.²⁻⁵ This is a case report of ectopic cervical tissue presenting as a cervical cystic swelling in a child.

Case Report. A 10-year old male child was referred to our hospital due to left-sided neck swelling of approximately 2 weeks duration. The swelling appeared suddenly and was not painful. There was no history of other complaints. Examination revealed a healthy looking child with a large 8 x 3 cm firm cystic swelling occupying the upper part of the left side of the neck. The swelling was not tender, and no other abnormalities were detected. Ultrasound of the neck revealed a multilocular cystic swelling of about 8 x 2 cm in size (Figure 1). This was confirmed by computerized tomography (CT) scan of the neck, which showed a

deep seated cystic swelling (Figure 2). The diagnosis of a neck lymphangioma was suspected, and the patient underwent surgical excision. During exploration there was a left sided cystic neck swelling that extended posteriorly to the spine. The swelling was excised totally and found to contain darkish brown fluid most likely altered blood. Microscopic examination revealed a thick cyst with a wall composed of dense fibrous tissue which contained abundant lymphocytes with lymphoid follicles and some clusters of epithelial cells, many cholesterol granulomas, and Hassell's corpuscles with thymic tissue. The lining was simple cuboidal or flattened epithelium and the wall showed some smaller cysts separated by fibrous septa. Post-operatively, the patient did well and was discharged home on the 2nd post-operative day.

Discussion. Embryologically, the thymus gland develops around the 6th week of intrauterine life from the ventral aspect of the 3rd pharyngeal pouch. It starts as paired primordia that elongate caudally to form the thymopharyngeal tracts. At about the 7th week of intrauterine life, these 2 tracts fuse incompletely in the midline to form the thymus gland which, and because of its attachment to the

From the Division of Pediatric Surgery, Department of Surgery, Qatif Central Hospital, Qatif, Kingdom of Saudi Arabia.

Received 9th January 2001. Accepted for publication in final form 8th April 2001.

Address correspondence and reprint request to: Dr. Ahmed H. Al-Salem, PO Box 18432, Qatif 31911, Kingdom of Saudi Arabia. Fax. +966 (3) 8360326. E-mail: asalem56@hotmail.com

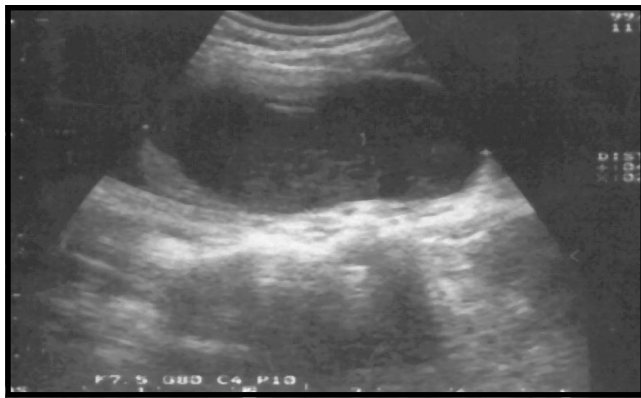


Figure 1 - Ultrasound of the neck showing a large cystic swelling of the neck.

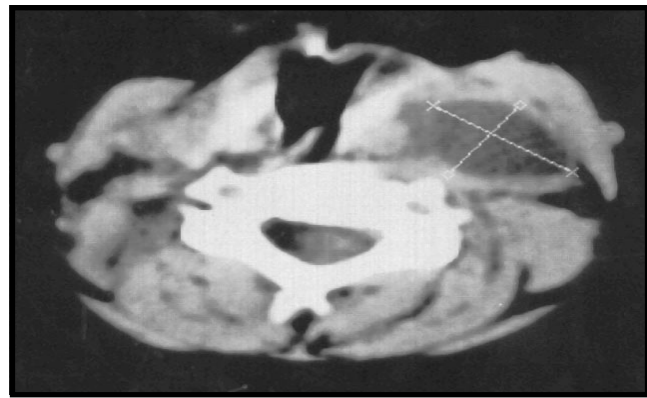


Figure 2 - Computerized tomography scan of the neck outlining the location of the cystic swelling which extended posteriorly to the spine.

pericardium, descends into the superior mediastinum to lie anterior to the pericardium and great vessels.^{1,3} As a result, thymic tissue may be seen along the line of descent of the thymopharyngeal tracts extending from the original position in the neck at the level of the hyoid bone to the final position in the superior mediastinum.² Noback, in an autopsy study of 65 random infants reported an 80% incidence of ectopic cervical thymic tissue.⁴ Fortunately, most thymic remnants in the neck remain dormant, and only very few of them enlarge to become visible or cause symptoms.

Ectopic thymic tissue is usually seen along the line of descent of the thymopharyngeal tracts, but it has also been identified in other sites including the base of the skull,⁶ pharynx,⁶ trachea,⁷ and posterior mediastinum.⁸ The presence of ectopic thymic tissue away from the line of descent of the thymopharyngeal tracts is most likely a result of migrational defects resulting in a loss and sequestration of part of the developing thymus gland and its migration with the local tissue to finally assume these abnormal ectopic positions.⁸ Whereas ectopic cervical thymic tissue, which is the most common, is usually asymptomatic and only apparent clinically, ectopic thymic tissue at other sites is difficult to recognize and may produce symptoms in the form of dysphagia or airway compression.^{5,9} Normally, the thymopharyngeal tracts become obliterated during the 7th and 8th week of intrauterine life. Persistence of these tracts with subsequent cystic dilation is the most likely explanation for the development of congenital thymic cysts.¹⁰ Thymic cysts at other sites, other than along the line of descent of the thymopharyngeal tracts arise most likely as a result of cystic degeneration of a solid ectopic thymic mass of tissue.⁵ Histologically, thymic cysts are easily differentiated from other congenital cysts, by the presence of thymic tissue with the pathognomonic

Hassell's corpuscles. Epithelial cells usually line the wall of the cyst and not uncommonly, cholesterol crystals as well as parathyroid tissue may be seen. Malignant transformation has been reported in thymic cysts in adults, but none so far in children.¹¹⁻¹³

Although ectopic thymic tissue is not uncommon, clinical congenital thymic cysts on the other hand are very rare. In 1982, Al-Shihabi and Jackson collected only 35 cases in the English literature.¹⁴ This can be explained by the fact that most thymic remnants in the neck remain dormant. The majority of thymic cysts in children are seen in the neck. Hendrickson et al in a review of 14 thymic cysts treated over a period of 13 years found 7 cervical, 5 mediastinal, and 2 extended from the neck into the mediastinum.⁹ Thymic cysts can be unilocular or multilocular and for unknown reasons they are more common on the left side and in males compared to females. Although the majority of thymic cysts are asymptomatic, not rarely they can present with life threatening symptoms. This is specially so with mediastinal cysts which tend to be larger in size.⁹ Hemorrhage into the cyst or a recent upper respiratory tract infection may lead to sudden enlargement of the cyst. Spontaneous hemorrhage into the cyst although rare may occur, and this was the most probable cause of the sudden appearance and enlargement of the cyst in our patient.

The differential diagnosis of cystic lesions in the neck in children usually includes lymphangioma, thyroglossal duct cyst, cleft cyst, dermoid cyst and teratoma, but because of their rarity, congenital thymic cysts are usually not considered. Add to this the similarity of their clinical and radiological features to the more common lesions. In fact in the majority of cases, the diagnosis is only made on histological examination. Congenital thymic cysts, although rare, should be included in the differential diagnosis of cervical and mediastinal cystic swellings in children. Physicians caring for these patients

should be aware of this and although the majority are asymptomatic, they can cause life threatening complications.

References

1. Moore KL. The developing human. Clinically oriented Embryology. 4th ed. Philadelphia(PA): WB Saunders; 1988. p. 179-184.
2. Gilmour JR. Some developmental abnormalities of the thymus and parathyroides. *J Path Bact* 1946; 52: 213-218.
3. Tovi F, Mares AJ. The aberrant cervical thymus: embryology, pathology and clinical implications. *Am J Surg* 1978; 136: 631-637.
4. Noback GJ. Contributions to topographic anatomy of the thymus gland with particular reference to its changes at birth and in period of newborn. *Am J Dis Child* 1921; 22: 120-144.
5. Spigland N, Bensoussan AL, Blanchard H, Russo P. Aberrant Cervical thymus in children: three case reports and review of the literature. *J Pediatr Surg* 1990; 25: 1196-1199.
6. Zarbo JR, McClatchey M, Areen RG, Baker SB. Thymopharyngeal duct cyst: A form of cervical thymus. *Ann Otol Rhinol Laryngol* 1983; 92: 284-209.
7. Lewis MR. Persistence of the thymus in the cervical area. *J Pediatr* 1962; 61: 887-893.
8. Al-Salem AH. Ectopic thymic tissue simulating a posterior mediastinal mass. *Eur J Pediatr Surg* 1992; 2: 106-107.
9. Hendrickson M, Azarow K, Ein S, Shandling B, Thorner P, Daneman A. Congenital thymic cysts in children - mostly misdiagnosed. *J Pediatr Surg* 1999; 33: 821-825.
10. Mikal S. Cervical thymic cyst: A case report and Review of the literature. *Arch Surg* 1974; 109: 558-562.
11. Babu MK, Nirmala V. Thymic carcinoma with glandular differentiation arising in a congenital thymic cyst. *J Surg Oncol* 1994; 57: 277-279.
12. Leong ASY, Brown JH. Malignant transformation in a thymic cyst. *Am J Surg Pathol* 1984; 8: 471-475.
13. Shu-ichi Y, Yamazaki H, Kato T, Babu MK, Nirmala V. Thymic Carcinoma which developed in a thymic cyst. *Internal Med* 1996; 35: 215-218.
14. Al-Shihabi M, Jackson J. Cervical thymic cyst: case report. *J Otolaryngol* 1992; 96: 181-189.